

The Extended Role of Total Laryngectomy in Patients with Severe Neuromuscular Disabilities

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ABSTRACT

Amongst the myriad of problems that patients with severe neurodisability encounter is that of recurrent airway obstructions and intractable aspiration pneumonias due to dysfunction of the protective mechanism of the larynx. Here, we describe the use of total laryngectomy as a surgical option for these patients. Case was 24 year old male with severe cerebral palsy suffering from recurrent episodes of airway obstruction and aspiration pneumonia. In the severely disabled patient with recurrent aspiration pneumonias, total laryngectomy may present as a viable option, especially in those patients who have been previously incapable of effective verbal communication.

Key words: Aspiration pneumonia, airway obstruction, neuromuscular disorder, laryngectomy

Şiddetli Nöromuskuler Bozukluğu Olan Hastalarda Total Larenjektominin Genişletilmiş Rolü

ÖZET

Şiddetli nörolojik bozukluğu olanlarda sayısız problemler arasında tekrarlayan hava yolu tıkanmaları ve larinks koruyucu mekanizma disfonksiyonu nedeniyle dirençli aspirasyon pnömonileri yer almaktadır. Burada, bu hastalar için cerrahi bir seçenek olarak total larenjektomi kullanımını tarif edilmektedir. Olgu hava yolu tıkanıklığı ve aspirasyon pnömonisi ataklarıyla gelen ciddi serebral palsili 24 yaşındaki erkek idi. Tekrarlayan aspirasyon pnömoniler ile gelen, özellikle etkili sözlü iletişimin daha önceden aciz olmuş ağır özürli hastalarda total larenjektomi görülebilir bir seçenek olarak sunulubilir.

Anahtar kelimeler: Aspirasyon pnömonisi, havayolu tıkanıklığı, nöromuskuler bozukluk, larenjektomi

INTRODUCTION

The larynx serves to protect the lower airway and facilitates respiration and phonation. Abnormalities or loss of its protective function place affected patients at a high risk of airway obstructive episodes and aspiration pneumonia (1). Permanent loss of this function, as seen in patients with neuromuscular disorders such as cerebral palsy and motor neurone disease, can result in intractable recurrent episodes which can significantly increase patients' morbidity and raise mortality rates up to as

high as 25% (2). Such patients are usually wholly dependent on family members and carers for their daily care, and in particular, care of their airway to prevent such episodes from occurring. This can often be challenging. In addition, there are the large psychological and financial burdens on family members and carers that repeated hospital admissions and specialised patient care can bring. On a larger scale, the estimated cost of treating a single episode of aspiration pneumonia with intravenous antibiotics on an intensive therapy unit in the United States averaged \$30,400 in 2008, a cost that is likely to

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be much higher in today's economic climate (3).

Surgical procedures designed to control intractable recurrent aspiration have been described in the literature, including glottic closure, cricopharyngeal myotomy, epiglottopexy, laryngotracheal separation, tracheostomy and total laryngectomy. We describe the case of a patient with severe cerebral palsy and intractable episodes of recurrent airway obstruction and aspiration pneumonia who underwent a total laryngectomy with good effect. Not only did the procedure solve the problem but had the added advantage of facilitating management by the patient's carers. The case for considering total laryngectomy in patients with severe neurodisabilities and the issues that surround it are discussed.

CASE

A 24-year old Caucasian male with severe cerebral palsy was referred to our department for management of recurrent episodes of airway obstruction due to the accumulation of thick, mucoid secretions and aspiration pneumonias, secondary to poor laryngeal function. In his recent history, he underwent seven acute admissions over a 2-month period and suffered an epileptic fit on one occasion due to obstructive episodes. He was otherwise non-verbal, blind and had musculoskeletal contractures associated with neck hyperextension. He was, however, able to recognise voice and touch. He had undergone a Nissen's fundoplication 20 years prior for gastro-oesophageal reflux disease (GORD) and was currently fed through a percutaneous endoscopic gastrostomy (PEG) tube for the last six years. Primarily, his foster mother cared him for.

Flexible nasoendoscopy revealed overall floppiness of the supraglottis with collapsing of the epiglottis and arytenoids during inspiration due to poor neuromuscular tone. There was evidence of both pseudobulbar and bulbar palsy. This combined with his hyperextended neck, prognathic jaw, bulky tongue base and thickened airway secretions were contributing to his frequent airway obstructions.

A best-interest multi-disciplinary meeting was held to discuss management options to reduce his morbidity. Among those present were his GP, respiratory physician, ENT surgeon, physiotherapists, occupational therapists, speech and language therapists, various nurse specialists and his family members. Multiple strategies were considered including the formation of a tracheostomy and total laryngectomy. After considering the pros and cons (as stated in Table) and with agreement from the court of protection, it was decided that the latter would provide the best quality of life outcome for the patient in his current state.

The procedure was performed without incident and the patient was transferred to the intensive therapy unit post-operatively where he remained for five days. He had an unremarkable recovery period and was discharged home on post-operative Day 21. Eighteen months later, he remains asymptomatic without any further obstructive episodes or hospital admissions. His carers have found that his daily care, and in particular, that of his airway had been much easier and that his quality of life and that of all involved in his daily care had dramatically improved (Figure 1).

Table. Pros and Cons of Tracheostomy vs Total Laryngectomy

	<i>Tracheostomy</i>	<i>Total Laryngectomy</i>
<i>Advantages</i>	<p><i>Technically less challenging hence quicker to perform</i></p> <p><i>Less laryngeal trauma</i></p> <p><i>Ideal for patients who are unfit for a larger operation</i></p> <p><i>Reversible</i></p>	<p><i>Technically more challenging, but with minimal complications in experienced hands</i></p> <p><i>A more definitive solution</i></p> <p><i>Minimal need for post-operative stoma care</i></p> <p><i>Ideal for patients with poor laryngeal function and poor wound healing</i></p> <p><i>Lesser chances of pharyngo-cutaneous fistula compared to when performed for cancers</i></p>
<i>Disadvantages</i>	<p><i>Loss of voice production</i></p> <p><i>Needs regular stoma care</i></p>	<p><i>Loss of voice production</i></p> <p><i>Irreversible</i></p>



Figure. View of stoma post laryngectomy

DISCUSSION

Patients with neuromuscular disorders are prone to impaired functioning of muscles due to either primary pathology of the muscle or indirectly via pathology of the central (pseudobulbar) and peripheral (bulbar) nervous systems. Common clinical features include muscular weakness, loss of muscular control, increased spasticity and myoclonus, all of which can affect the larynx and its surrounding muscles as observed in our case (4). Many surgical options have been described to treat and prevent intractable aspiration pneumonia although there have been none specifically designed to control chronic airway obstruction episodes. In 1975, Montgomery advocated reversible simple glottic closure. Unfortunately, none of his 12 patients had sufficient recovery of laryngeal sphincteric function to warrant an attempt at reversal and dehiscence of the closure was found to be a common complication, necessitating a more definitive solution (5). Cricopharyngeal myotomy was later suggested as a simple management strategy by Loizou. et al (1980) for patients with dysphagia secondary to motor neuron disease. Although there were some initial improvements in swallowing and hence reduction in episodes of aspiration pneumonia, the progressive nature of the disease resulted in recurrence of these symptoms in later years (1). In 1983, Brookes et al described epiglottoplexy, which involved the epiglottis being flapped posteriorly and fixed onto the aryepiglottic fold. Although the author reported good post-operative outcomes and near normal voice preservation, there were later cases of continued salivary aspiration particularly in patients with poor healing capacities (1,6).

Laryngotracheal separation, on the other hand, involves

closure of the larynx, with or without anastomosis to the oesophagus, and the creation of a permanent tracheostomy at the level of the first or second tracheal ring. Gelfand et al reported good outcomes with the procedure and minimal post-operative morbidity (7). However, in addition to requiring post-operative stoma care, these patients have high rates of fistula formation, occurring in as many as 1 in 3 patients. This can be reduced with the use of strap muscle flap-reinforced imbrication closure of the proximal tracheal stump (8).

Elective tracheostomy is a more popular surgical option whereas a total laryngectomy is rarely done but has been described with good effect for patients with neuromuscular disabilities and was hence suggested as a long-term solution for our patient. A tracheostomy is an artificially created opening into the trachea via the neck and is held patent with a tube (9). It is described as technically less complex with less surgical trauma to the neck in comparison to a total laryngectomy and has the added advantage of a reversal (9). Common complications include persistent bacterial colonisation, accidental dislodgement and obstruction of the tracheal tube (9). As a result, close monitoring is essential in ensuring the temporary tracheal tract remains patent during dislodgement. In addition, stoma care (regular humidification, suction and cleaning) is a vital part of the post-operative management which requires dedicated time and training of the carers (9).

In comparison, total laryngectomy involves the removal of the larynx, hence permanently separating the upper airway and alimentary tract (Figure 1). It is a procedure traditionally performed for laryngeal cancers incurable by chemoradiotherapy or laser resection. As it involves the loss of speech function, it is usually considered when all other treatment options are not indicated or have failed. It has been advocated as the procedure of choice in selected cases of intractable aspiration, particularly in patients with poor recovery of laryngeal function or in cases of compromised wound healing. It benefits from a short procedural time and can potentially be performed under local anaesthetic (1). Also, because the tissues of the neck are relatively healthier than in patients with carcinoma who have potentially been exposed to radiotherapy, healing is far more superior and patients are less likely to suffer from complications such as wound breakdown and fistula formation. The main disadvantage is that it is irreversible, though with careful patient selection, this rarely becomes an issue.

A common disadvantage for both tracheostomy and total laryngectomy is the loss of voice (Table). Although many patients with neuromuscular disorders may not have or could lose their speech function as a result of the debilitating nature of their disease, their basic voice production may be the only form of communication for their loved ones, more notably so in children. The loss of voice through surgical intervention may have a huge psychological impact on family dynamics. However, Takano et al. have shown that both procedures not only improved the underlying depression and mood of patients and their families but also the feeding status and clinical outlook (10). Aphonia must thus be weighed against these factors and discussed with the family and carers on an individual patient basis.

The importance of a multi-disciplinary approach in managing these patients must not be underestimated. As some patients with neurodisability may not have the capacity to make decisions or consent (especially those being considered for surgical intervention), the responsibility may fall on the primary clinician to decide on a patient's best healthcare interest. It is thus vital that these decisions are made alongside family members and close relatives (including those with Lasting Power of Attorney), carers and other healthcare professionals involved, both in primary and secondary care, to ensure the provision of sufficient information and the continuity of care. In some cases, an Independent Mental Capacity Advocate (IMCA) or Court of Protection order may be necessary to proceed.

In conclusion, the larynx plays a large role in protection of the lower airway. Patient with severe neuromuscular disability could lose this protective function which predisposes them to recurrent aspiration pneumonia and airway obstruction. A total laryngectomy may be a viable management option in a selection of these patients. Such a decision should be discussed in a multi-disciplinary environment with the family members and carers to weigh the benefits and risks

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